ACUTE LYMPHOBLASTIC LEUKEMIA PRESENTING WITH SKIN RASH AND FOREHEAD SWELLING IN A 31-YEAR-OLD MALE

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Abstract

Leukemia cutis is the infiltration of neoplastic leukocytes or their precursors into the epidermis, the dermis, or the subcutis of patients with leukemia, resulting in clinically identifiable cutaneous lesions. It is especially rare as the presenting symptom of leukemia and is associated with a poor prognosis. We present a case of acute lymphoblastic leukemia, whose initial complaints were skin lesions and forehead swelling, which we believe to be features of leukemia cutis.

Keywords: *acute lymphoblastic leukemia, skin rash, leukemia cutis, low back pain*

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Introduction

Leukemias are malignant disorders of the white cell compartment, characteristically associated with increased numbers of white cells in the bone marrow and/or peripheral blood. Leukemia cutis is the infiltration of neoplastic leukocytes or their precursors into the epidermis, the dermis, or the subcutis of patients with leukemia, resulting in clinically identifiable cutaneous lesions.[1] It is a rare condition that mainly described in case reports or small case series. Although more commonly seen in acute leukemias of myeloid and monocytic lineage, lymphoblastic leukemias can also involve the skin.[2] A diagnosis of leukemia cutis generally portends a poor prognosis and strongly correlates with additional sites of extramedullary involvement.[1] The diagnosis is difficult in the absence of a systemic presentation of acute leukemia. Therefore, when cutaneous expression of the disease is the presenting symptom, it is frequently misdiagnosed. It can present anywhere on the skin, with variable morphological features including macules, papules, plaques, nodules, masses and ulcers, which makes it difficult to clinically distinguish from other non-specific skin lesions.[3]

Case report

A 31-year-old male was admitted in the department of medicine of Dhaka Medical College Hospital with multiple violaceous nodules, papules and plaques over both lower limbs for 2 months. The lesions were purple to violet in color, non-tender and non-itchy. He noticed a swelling on the left side of forehead 2 months back, which was non-painful, and gradually increasing in size. There was no history of trauma prior to the onset of this swelling.

He also had been suffering from low back pain for the last 15 days, which was initially mild but later severe, worsened during activity and caused sleeping difficulties. It was non-radiating and not associated with any morning stiffness. He also complained of shortness of breath for the last 10 days, which was progressively increasing,

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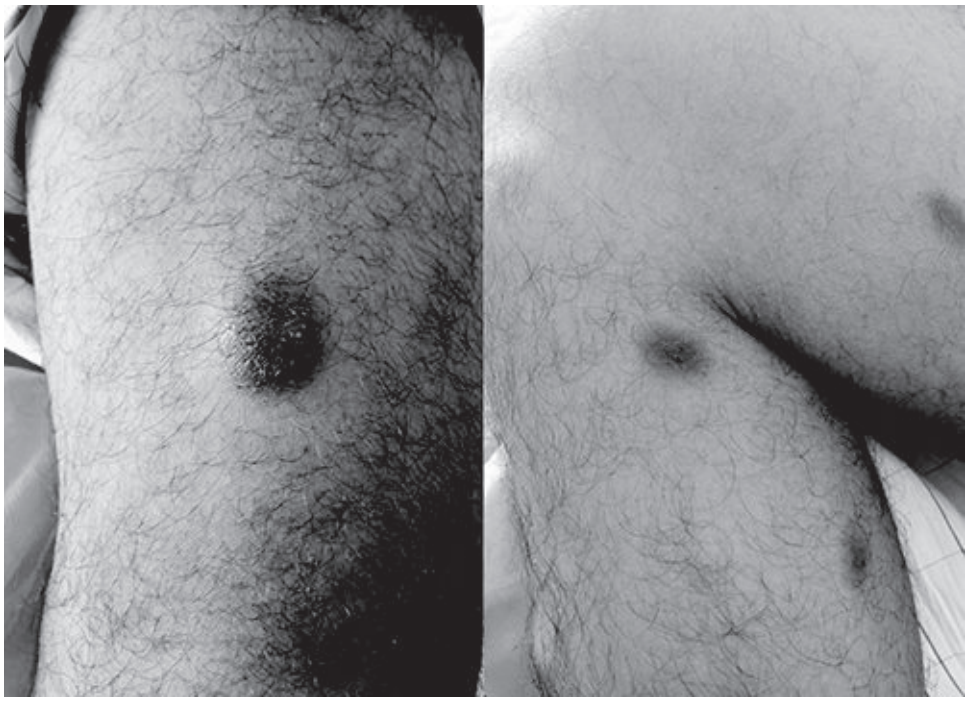

Figure 1: *Multiple violaceous nodules, papules and plaques over left (A) and right (B) lower limbs.*

more marked during moderate to severe exertion and not associated with any cough, orthopnea or paroxysmal nocturnal dyspnea.

On further query, he mentioned of occasional episodes of fever over last 1 month, which was low grade, intermittent, with a maximum recorded temperature of 100°F, subsided by intake of paracetamol with sweating, not associated with chills & rigor or evening rise of temperature. On subsequent query, he also mentioned of mild degree of hearing loss of sudden onset, affecting both ears for 15 days, slowly progressive and accompanied by tinnitus. There was also history of significant weight loss over last month, which was unintentional and evident by loosening of clothes & shoes.

On general examination, the patient was anxious and mildly anemic. There were multiple violaceous nodules, papules and plaques over both lower limbs, which were non-pruritic, non-tender and not blanching on pressure. There was a swelling on the left side of forehead which was approximately 3x3 cm in size, globular in shape, firm in consistency, non-tender, non-fluctuating, fixed with underlying structure but not fixed with overlying skin. Respiratory system examination revealed left sided pleural effusion. Neurological examination revealed bilateral conductive deafness of the 8th cranial nerve. Examination of rest of the systems revealed no abnormality.

Figure 2: *A swelling over the left side of forehead, approximately 3x3 cm in size, globular in shape, firm in consistency, non-tender, non-fluctuating, fixed with underlying structure but not fixed with overlying skin.*

Initial CBC showed: Hb – 9.2 g/dl, WBC – $13x10^9$/L, Neutrophil – 80%, Platelet – $103x10^9$/L, ESR – 25 mm in 1st hour. PBF showed microcytic hypochromic anemia with thrombocytopenia with leukocytosis. Chest X-ray revealed left sided pleural effusion. In urine R/E, pus cell count was 6-8/HPF. CRP was 33.74 mg/L. Pleural fluid study revealed exudative fluid, ADA was 18.2 U/L and gene-xpert was negative. USG of whole abdomen showed G-I fatty liver and left sided pleural effusion with adjacent basal consolidation. CT scan of chest showed nothing other than left sided pleural effusion. CT scan of head showed soft tissue swelling at left frontal region of the scalp. Pure tone audiogram revealed mild conductive hearing loss in right ear and low frequency hearing loss in left ear. Biopsy of skin lesions showed non-specific findings. CBC was repeated after 2 days which showed Hb – 9.4 g/dl, WBC – $11x10^9$/L, Platelet – $110x10^9$/L, ESR – 20 mm in 1st hour.

X-ray lumbosacral spine was normal. OGTT, S. creatinine, uric acid, calcium, albumin, PO_4, PTH and LDH levels were normal. S. bilirubin, ALT, AST, ALP were also normal. HBsAg, Anti-HCV, Anti-HIV 1 & 2 were negative. Rheumatoid factor, Anti-CCP, ANA, Anti-ds-DNA, p ANCA and c ANCA were negative. S. ACE level was 44 U/L. In induced sputum, AFB was not found and gene-xpert was negative. At this point, we could not reach a definite diagnosis.

After 7 days, we again ordered CBC which showed Hb – 10 g/dl, WBC – $29x10^9$/L, Lymphocyte – 54%, Platelet – $80x10^9$/L, ESR – 10 mm in 1st hour with 30% atypical cells. With these findings, we had to repeat PBF again which showed: RBC - normocytic normochromic, WBC - Increased with left shifted, majority cells are small to medium sized atypical mononuclear cell having scanty cytoplasm, high N:C ratio, coarse chromatin & inconspicuous nucleoli resembling lymphoblast, Platelet - reduced. These features were suggestive of acute lymphoblastic leukemia. Bone marrow study was then done and the report showed hypercellular marrow, increased myeloid erythoid ratio, depressed but normoblastic erythropoiesis, markedly depressed granulopoiesis with maturing into segmented forms and normal megakaryocytes. It also showed markedly increased lymphoid cells with shift to the left, heterogenous population of blasts (> 90%) having scanty cytoplasm, high N:C ratio, coarse chromatin, inconspicuous nucleoli resembling lymphoblast. These features were also suggestive of acute lymphoblastic leukemia.

With the diagnosis of acute lymphoblastic leukemia, we transferred the patient to the Hematology department for further management.

Discussion

Typically hematological malignancy presents in the form of pale skin and mucous membrane, weakness, fever, bruising, bleeding, bone pain, abdominal pain or lymphadenopathy.[4] Leukemia is a malignant neoplasm of the hematopoietic system. Cutaneous involvement with acute leukemia is fairly uncommon and usually a relatively late event. Leukemia cutis clinically demonstrates skin infiltration by leukemia and can present as macules, papules, plaques, nodules, masses and ulcers.[3] In our patient, it presented as nodules, papules and plaques over lower limbs, and mass like soft tissue swelling on forehead.

Low back pain is common in general population and has various causes. Leukemia should be considered in patients with unexplained pain in the back or of the epiphysis of the long bones, or joint pain out of proportion to the severity of existing arthritis when there is no history of trauma.[5] Bone pain in leukemia may be due to leukemic infiltration of marrow cavity or periosteum. Other features of leukemic infiltration in our patient are pleural effusion, hearing loss and tinnitus.

We herein described a patient with ALL who presented with leukemia cutis as initial manifestation. This report represents one of the few cases of ALL presenting with leukemia cutis.

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